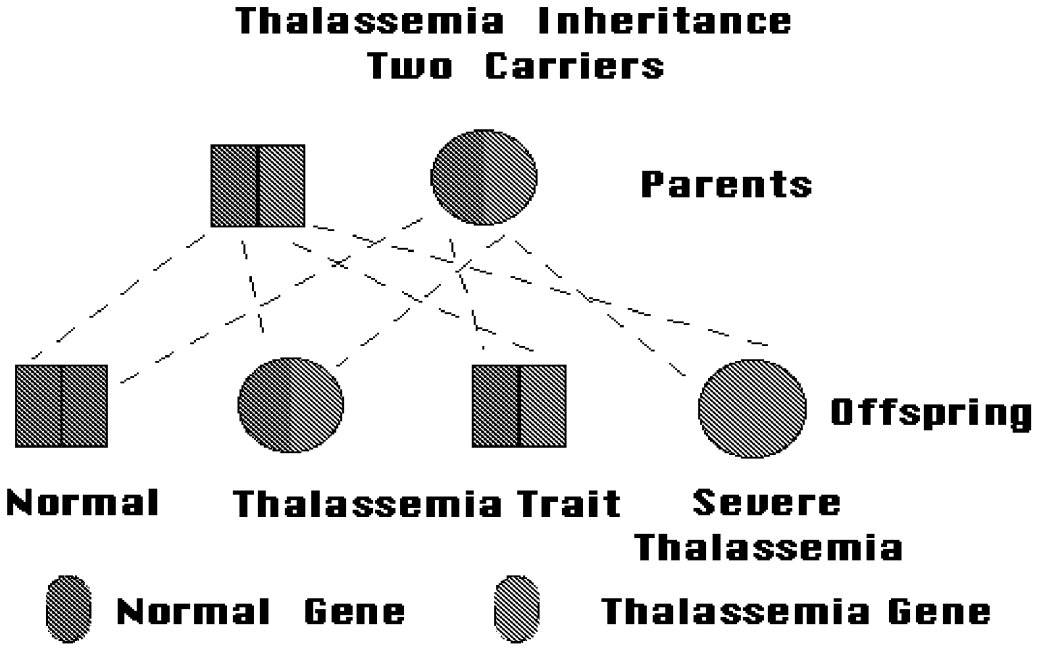
Treatment of β-Thalassemia with Chinese Herbs

by Subhuti Dharmananda, Ph.D., Director, Institute for Traditional Medicine, Portland, Oregon

β-**thalassemia** is a rare blood disorder in which hemoglobin is improperly formed due to an inherited genetic defect (mutation). The beta chain of the hemoglobin molecule is affected, which contributes part of the name of this disease (there is also an a-**thalassemia**). The disease is most prevalent in the Mediterranean region; and thalassa is Greek for sea, indicating the Mediterranean Sea in this context (-emia indicates the disorder is of the blood). The disease has been called Mediterranean anemia, since the primary effect is a severe anemia. The disease becomes apparent a few months after birth and, depending on its severity (for example, whether one or both genes contributed by the parents are affected), the anemia can be fatal within the first few years of life. For those with one gene affected, the disease is called β-**thalassemia** minor; when both genes are affected, the disease is called β-**thalassemia** major.



Severe β-**thalassemia** (major) occurs when both parents are carriers of the defect in the hemoglobin beta chain (illustrated right); carriers can survive to adulthood, but suffer from chronic anemia and secondary effects of it.

The disease is rare in China, but has been noted, particularly in the Nanning of Guangxi Province. Even though the Mediterranean Sea is the central area where the disorder occurs, it spreads eastward through the middle East and into East Asia. It is likely that the disease was transmitted over thousands of years by the caravans that traded between the Mediterranean area (Europe and Africa) and the populations to the East.

Modern treatments include blood transfusions, treatment with hydroxyurea (which promotes hemoglobin production), and stem cell transplants. Genetic engineering to repair the defective gene is a hoped for solution in the coming decades. Its treatment with Chinese medicine is of some general interest, in that it may reflect helpful measures that can be applied to a number of blood disorders, such as sickle cell anemia, that involve genetically-based defects in hemoglobin and resulting impairment of the blood.

According to the Chinese traditional descriptions, the "kidney" system (*shen*) is the source of the bone marrow (*sui*) that produces blood cells. The kidney and marrow store the essence that is present at birth and that is affected by prenatal factors (e.g., genetics). Tonifying the kidney essence is the basic principle of therapy to be applied. The herbal therapy has been called *Bushen Shengxue Fang* (*bushen* = tonify the kidney; *shengxue* = generate blood; *fang* = recipe) or, alternatively, *Busui Shengxue Fang*, or *Yisui Shengxue Ling* (*yisui* = to boost up the marrow; *ling* = effective **remedy**). The recipes used for this therapy can vary; they have not been identified fully in Chinese publications. One of the formulations that has been revealed is (1):

Astragalus	24 g
Lycium fruit	15 g
Antler gelatin	12 g
Tortoise shell gelatin	12 g
Donkey hide gelatin	12 g
Tang-kuei	12 g
Peony	12 g
Rehmannia, cooked	12 g
Ho-shou-wu	12 g
Placenta	12 g
Ginseng	10 g
Baked Licorice	6 g

The herb amounts in grams represent the quantity used to make a one day dose by decoction, which is cooked with 40 grams of magnetite powder (natural source of iron). This formulation is used in treating aplastic anemia and idiosyncratic thrombocytopenia, but may be applicable to other marrow disorders; presumably, it could be used for sickle cell anemia patients as well.

Since 1980, researchers at the Guang'anmen Hospital of the China Academy of Traditional Chinese Medicine (Beijing) and at the Hematology Institute of the Chinese Academy of Medical Sciences have been investigating treatment of β-**thalassemia**. In the first formal report from this group (2), they described administration of a capsule containing rehmannia, tortoise shell gelatin, donkey hide gelatin, cornus, and other herbs to children (ages 5-14 years) with β-**thalassemia**. The dosage was 3-4 capsules at one time, 3 times daily, with 2 months as a course of therapy. The authors reported improvement in hemoglobin levels and, through studies of RNA, suggested that transcription of RNA was enhanced and the gene defect was somewhat compensated for. The therapy was said to be effective for those with β-**thalassemia** minor, but not for those with β-**thalassemia** major.

In a follow-up study (3), with 17 patients having β-**thalassemia** minor treated during 3 months starting October 1998, this grouped described the therapy as containing 12 ingredients, including cornus and millettia. They concluded that 16 cases were improved: the symptoms were ameliorated; hepatosplenomegaly decreased; hemoglobin was significantly elevated; and no side effects were observed.

This group of researchers gave further information on this subject at the 2000 International Congress on Traditional Medicine in Beijing (4). They concluded that the positive effect of the herbs on this disease confirmed the TCM theory that the kidney nourishes the marrow, and emphasized that the mechanism of action was at the level of the DNA, "unlocking the gamma-gene, stimulating mRNA expression of gamma-globin, and inducing synthesis of hemoglobin to compensate for the defect of the beta-gene."

REFERENCES

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